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Excessive Daytime Sleepiness in Behçet's Disease with Diencephalic Lesions and Hypocretin Dysfunction

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Key Words

Excessive daytime sleepiness • Hypersomnia • Behçet's disease • Hypocretin

A 31-year-old man with Behçet's disease suffered from acute diplopia, hypersomnia (>12 h sleep/day) and sleepiness (Epworth sleepiness scale: 14 points). Cranial MRI revealed diencephalic

lesions with left-sided subthalamic gadolinium enhancement (arrow; fig. 1A–C). Wake-promoting hypothalamic hypocretin-1 in the cerebrospinal fluid (CSF) was decreased (215 pg/ml; normal: >320 pg/ml) [1]. Following treatment with prednisone for 1 month and with azathioprine, diplopia, sleepiness and hypersomnia disappeared within 2 months. Hyperintense lesions vanished on cranial MRI (fig. 1D). CSF studies revealed a normal hypocretin-1 level (400 pg/ml). This is the first reported patient with Behçet's disease with a low and treatment-responsive CSF hypocretin-1 level.

Reference

- 1 Baumann CR, Bassetti CL: Hypocretins (orexins) and sleep-wake disorders. *Lancet Neurol* 2005;4:673–682.

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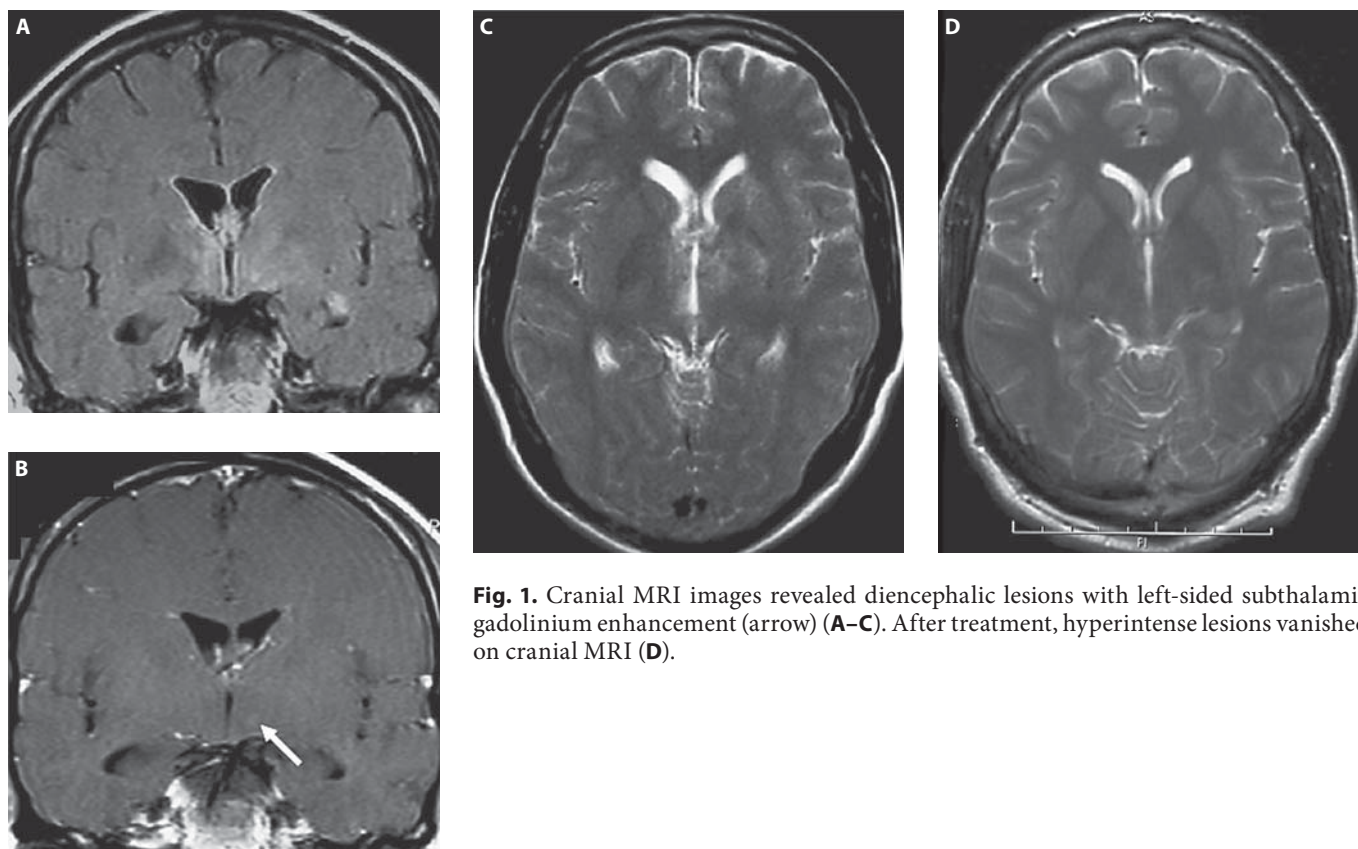


Fig. 1. Cranial MRI images revealed diencephalic lesions with left-sided subthalamic gadolinium enhancement (arrow) (A–C). After treatment, hyperintense lesions vanished on cranial MRI (D).